Ms Anne Recordon
Physiotherapist
Neuro Rehab Results
Queenstown

Saturday, August 12, 2017
14:00 - 14:55  WS #106: Managing Dystonia in Children and Adults
15:05 - 16:00  WS #117: Managing Dystonia in Children and Adults
(Repeated)
Dystonia in Children and Adults

Anne Recordon
anne@neurorehab.co.nz
0274934819
Saturday Room 9
2pm-3pm
3pm-4pm repeat
GPCME workshop
Childhood dystonia
and Focal / Cervical
Dystonia
2017 Christchurch
Anne Recordon Dip Phys, ADP, MHSc, NZRPT
anne@neurorehab.co.nz
Role of physiotherapy in dystonia

PLAN OF ATTACK

• The Brain and Basal Ganglia
• Physiotherapy in Children with dystonia
• Physiotherapy in Adults with dystonia
• Head movements and the muscles
• Adult Assessment/Intervention strategies
Abnormal tone and movement patterns

Site of the lesion - Basal Ganglia Nuclei
- Complex organisation of loops (neuronal networks or circuits)
- Anatomical somatotopic representation
- Neurotransmitter substance/somatotopic cells/ maladaptive neuroplastic changes
- Role BG Automatic program memorized embedded in relation to task i.e. activity dependent
- Loss of resultant feedforward motor plan ie purposeful pattern movement / task dependent muscle synergies
- Involuntary movement pattern initiated (dystonic spasms)

Kandel, Schwartz & Jessell 2001
Motor loops showing somatotopic subdivisions projecting to supplementary motor area, pre motor and motor cortex.

Involuntary initiation of dysfunctional motor programming planning and execution.

Intervention:
Retraining movement/challenging sensory motor input.
Use cognitive, visual cuing ....patterns to hold head up in the middle to use eyes for..?
Anticipatory Postural Adjustment (APA)

**Task/Motor Plan**

### Action demands
- **Mobility commands to joints and muscles for activity**

### Postural commands
- **Stability commands to joints and muscles**

**Parameter**
- **Focal action**
- **Anticipatory Postural Activity**

Fig 2.2 p13 Postural Control 2008 M Hadders Algra Eva Brogren Carlberg
Adult Focal/Cervical Dystonia definitions

“Cervical dystonia (the most common focal dystonia, frequently results in cervical pain and disability as well as impairments affecting postural control”
B E Crowner 2007

“Cervical dystonia has been described as involuntary twisting and turning of the neck caused by abnormal involuntary muscle contraction
Fahn, Marsden, Calne 1987
Definition: Dystonia

- Dystonia is classified as a movement disorder characterized by sustained or intermittent, involuntary muscle contractions causing twisting, repetitive movements, or abnormal postures.

- Dystonia can be focal (affect a specific area of the body), segmental (spread to two or more adjacent body regions), multifocal (involving two or more noncontiguous body regions), or generalized (involving a majority of the body).

- The multiple etiologies of dystonia include inherited (or genetic in origin), acquired, and idiopathic (or unknown cause).

- Focal dystonia is the most common form of dystonia and can involve any body part including the neck (cervical dystonia [CD]), limbs (limb dystonia), hands (focal hand dystonia), eyes (blepharospasm), mouth (oromandibular dystonia), or trunk (camptocormia).

Truong D 2005
Dystonia in children

• “A movement disorder in which involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures or both”

• Clinical Features:
  • Postures repeated within same child
  • Similar dystonic postures seen within children with dystonia
  • Triggered by attempts to move
  • Involuntary postures decrease or absent during sleep
  • “Overflow” may be present
  • Frequently seen in Dyskinetic CP with injury to basal ganglia thalamus or cerebellum

Darcy Fehlings 2010
Figure 2: Hierarchical classification tree of cerebral palsy sub-types.

Is there persisting increased muscle tone in one or more limbs?

Yes  

Are both sides of the body involved?

Yes  

Spastic bilateral CP

No  

Spastic unilateral CP

No  

Dykinetic CP

Is the tone varying?

Yes  

Reduced activity: tone increased

Dystonic CP

Increased activity: tone decreased

Choreo-athetotic CP

No  

Is there a generalized hypotonia with signs of ataxia?

Yes  

Ataxic CP

No  

Non-classifiable
Prevalence of CP by CP Type: Swedish study

Himmelmann K, Hagburg G, Uvebrant P. 2010
Impairments Taxonomy

Hypertonic
- Spasticity
- Dystonia
- Rigidity

Hyperkinetic
- Chorea
- Dystonia
- Athetosis
- Myoclonus
- Tremor
- Tics
- Stereotypes

Negative signs
- Weakness
- Selective motor control (SMC)
- Ataxia
- Dyspraxia
- Bradykinesia
- Balance
Clinical features with children with dystonic movement disorder (CP)

• Loss of head and trunk control with spasms pulling ...
• Affecting feeding breathing communication
• Unable to sit stand or sequence up against gravity
• Loss postural control interfered with by the spasms
• Affects both balance strategies and voluntary task related movements i.e intent initiates spasms
• i.e. poor postural stability and involuntary movements interfering with the planned task related movements
Impact of positioning

Overtime contractures and deformities
Elizabetha

I am still and in the middle I can hold my head up and use my eyes to learn
Video: Children with dystonia
Dystonia arms and mouth
Adult Focal/ Cervical dystonia

- Prevalence 4.98/100,000 in Europe
- Male-to-female ratio of 1 : 1.2.
- Peak age of onset is around 41.8 years
- Most cases are idiopathic
- Diagnosis on average takes 4 different specialists

Associated impairments
- Considerable impairment of ADL’s
- Pain and mood disorders in 41%

Reversa et al 2015
Assessment tools

• The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS)
• Visual Analogue Scale (VAS) for Pain.
• Cranio cervical Dystonia Questionnaire (CDQ-24)
• Disease severity scales such as the Tsui scale and the Cervical Dystonia Severity Scale
• Short Form Health Survey-36 (SF-36)
• cervical range of motion
TWSTRS Torticollis

Severity Scale (maximum = 35)

A. Maximal Excursion
Rotation (turn: right or left) 0 = None [0°]
1 = Slight [< 1/4 range, 1°–22°]
2 = Mild [1/4– 1/2 range, 23°–45°]
3 = Moderate [1/2– 3/4 range, 46°–67°]
4 = Severe [>3/4 range, 68°–90°]

Laterocollis (tilt: right or left, exclude shoulder elevation)
0 = None [0°]
1 = Mild [1°–15°]
2 = Moderate [16°–35°]

B. Duration Factor
C. Effect of sensory tricks
D. Shoulder elevation / anterior displacement
E. Range of motion
F. Time
TWSTRS Torticollis

Disability Scale (maximum 30)
- Work
- Activities of daily Living
- Reading
- Driving
- TV
- Activities outside the home

Pain Scale (Maximum 20)
- Severity of pain
- Duration of pain
- Disability due to pain

Injection record
Main muscle groups involved
Head movements

Directions and range
1. Head flexes 0-45°’s
2. Head extends 0-45°’s
3. Rotates to right 0-75°’s
4. Rotates to left
5. Side flexes to right 0-45°’s
6. Side flexes to left

Combination of muscle patterns
• Look at alignment and asymmetry...how head is being pulled by spasm
• Overuse causes pain
• Muscle imbalance produced by asymmetry and non alignment of head produces pain
Head flexion and rotation

**SCM**

*Sternocleidomastoid*

Right SCM causes neck rotation and chin tuck to right

Palpate

Opposite muscle will be lengthened and weakened

Anterocollis head pulled forward by sternocleidomastoics

Laterocollis head pulled to one side with rotation
Shoulder Shrugging

Trapezius (a) +
Levator scapulii (c)

Shoulder elevation is a common feature
Extension and rotation (retrocollis)

- Small deep muscles (postural) splenius capitus, cervicus, thoracis (fdj)
- Semispinalis (hij)

Side Flexion (ear to shoulder) (laterocollis)

- Splenius capitus (c)
- Scalenes and the Sternocleidomastoid
Cervical dystonia

Interferes with what function?
Intervention: Treatment of choice
Botox + Physiotherapy

• Limited research to review. Level of evidence level 11
• European Guidelines
• The team
• Quieroz: 4 weeks, 5 days, 75 mins per session  15 days post botox
• Reversa 2015: Oral medications may be used as adjunctive therapy to BoNT injection for symptomatic relief and include anticholinergic agents, baclofen, muscle relaxants, and benzodiazepines
• DBS with electrodes placed in the globus pallidus interna has been effective in the treatment of generalized dystonia; inconsistent results with the use of DBS for CD.
• Pisani 2017  oral medications poorly tolerated side effects. Trihexyphenidyl and bendiazepans
FES

Electrical stimulations were synchronized (FES-Sync), with two channels.

Parameters:
- Frequency 50 Hz;
- On-time 10 s, off-time 30 s (1:3 on-off ratio);
- Rise and decay times 2 s and 1 s, respectively;
- Pulse 200 μs.

Each FES treatment 30 minutes.

• A multimodal physiotherapy program can consist of active exercises, stretching, massage, relaxation, active and passive mobilization of the cervical spine, EMG biofeedback, or electrical stimulation of antagonist muscles.

Queiroz et al 2012
Practical session – let’s get moving

- **Stretches neck**
  - Flexors (SCM)
  - Rotators (SCM)
  - Side flexors
  - Deep extensors

- **Strengthening of opposing muscles**

- **Awareness of Posture and alignment during function**
  - Lower back
  - Check “chin in”
Planning to move in a different way

• “Tricks”
  • Sitting
  • Use of hands together
  • Holding chair
  • Elbows on table
• Increasing awareness of posture
• Management holistic

Lifestyle balance
• Meditation
• Yoga pilates?

• Daily activity own opiates and happy drugs
Case study

- “S” 3 year history
- Receiving botox
- School teacher
- Young child
- Symptoms began a year ago dropped when drinking
- Marked flexion head with rotation to left
- Shoulder elevation
- Diazepam 1mg and Botox A 75mg sternomastoid and 25 units levator scapula
Training Program

- Teaching standing
- Teaching desk work
- Family meals drinking and eating
- Rest positions
- Stretches
- Complementary medicine massage, spa pool
Strengthening

- Strengthening
- +FES
- Stretches
- Postural control mirror feedback
How do we learn to move with more efficiency?

- Neural plasticity
- Skill acquisition
- STM
- LTM

= learning

- to be in the middle
- to look / learn teach
- to hear / taste eat
- to communicate

Muscle Imbalance

Strong overused agonists

Weak underused antagonists

PAIN STIFFNESS

Observation and analysis of overactive muscles

Stretching And strengthening

Observe posture Use movement control strategies

Education positioning and alignment

BOTOX

+ 

PT

TEAM SLT OT PT

MEDS
Complementary medicine

• What have you found useful?
References


• Crowner B E. Cervical Dystonia: Disease Profile and Clinical Management. Physical therapy 2007; 87: 1511-1526

